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CONGENITAL DERMAL SINUSES: A SOURCE OF SPINAL MENINGEAL INFECTION AND SUBDURAL ABSCESSES.[1]

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CONGENITAL DERMAL SINUSES.

SACRAL and coccygeal pilonidal cysts or sinuses are common congenital anomalies, but they rarely extend to involve the meninges or the spinal cord. In higher spinal segments, however, sinuses, histologically identical with those occurring in the sacral and coccygeal regions, are found reaching and even penetrating the dura mater. Such congenital malformations, forming an epithelial tube from the skin of the back to the coverings of the spinal cord, are a potential pathway for infection of the subdural and subarachnoid spaces. This source of meningeal infection has been almost completely overlooked in medical literature, and yet once recognized and the condition looked for such a mode of infection is not as rare as might be assumed from the lack of reported cases.

Case 1.—L. I., a girl, aged 5, was referred to the University of Chicago Clinics from the Children's Memorial Hospital, Chicago. From her birth the patient's parents had noted a small dimple in the midline of the upper portion of the child's back. In all other respects the child seemed perfectly normal At 6 months of age the site of the dimple became swollen, purple in colour and discharged a slightly bloody material. The patient at that time had a high fever and sore throat. Shortly afterwards she began to vomit frequently. A physician noted that the back was rigid and on lumbar puncture found 150 cells in the spinal fluid. After standing for an hour a pellicle formed in the fluid. A portion was injected into a guinea-pig and the animal died three weeks later. Examination of the animal is said to have revealed evidence of tuberculosis. Another observer saw the patient at that time and found her "irritable and cranky." The fontanelle was not bulging; there was no strabismus and no Kernig's sign, but there was definite rigidity of the neck. He thought the child was suffering from an encephalitis. Subsequently another specimen of spinal fluid was injected into a guinea-pig and again the animal died. This was considered conclusive proof that the child was suffering from tuberculous meningitis.

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This illness, with great irritability, evening fever, vomiting, loss of appetite and rigidity of the back, continued with varying degrees of intensity for about eighteen months when, for no apparent reason, the child recovered and remained perfectly well until 5 years of age. During the intervening three years, however, the mother had noticed an occasional discharge from the dimple of a few drops of milky material, which was at times blood stained. This occurred two or three times a year and on two occasions the skin surrounding the dimple became red and swollen.

During November, 1932, the patient began to complain of pain in the neck and back, particularly at night. She was taken to the Children's Memorial Hospital. Examination at that time showed a spasm of the left sterno-cleidomastoid muscle, a scoliosis of the upper thoracic vertebræ with the convexity to the right and a small, slightly elevated dimple just above the spine of the 5th thoracic vertebra. Both knee and ankle jerks were very lively and a bilateral, sustained ankle clonus was present. The right plantar reflex was extensor and the left flexor. Roentgenograms showed an incomplete fusion of the laminæ of the 4th thoracic vertebra and a marked spinal curvature. After ten days in the hospital the patient's condition improved greatly and she was discharged. During the following months she was examined at frequent intervals in the out-patient department of that hospital. Her gait became progressively more spastic, the reflexes of the lower extremities more active, and at times local tenderness was present over the 4th and 5th thoracic vertebræ.

Because of this steady progression of her condition, she was readmitted to the hospital on April 22, 1933, for lumbar puncture. The fluid was clear, under normal pressure, contained three lymphocytes and gave a negative reaction for globulin (Pandy). The Wassermann test on this fluid was negative. She was again treated in the out-patient department. Late in June the patient began to scratch and rub the ulnar surfaces of both hands and arms, presumably as a result of paræsthesiæ. Due to this rubbing, it was noted on her next visit on July 3 that the ulnar surfaces of both forearms were red and inflamed. The findings on neurological examination at that time were essentially as they had been previously. Roentgenograms showed no change in the appearance of the thoracic vertebræ, or of the defect in the 4th vertebra. A lumbar puncture revealed a xanthochromic fluid under very low pressure, containing fifty red blood corpuscles and an increased quantity of globulin as tested by Pandy's method. Queckenstedt's test revealed a partial block in the spinal subarachnoid space.

On July 18, 1933, she was admitted to Bobs Roberts Hospital of the University of Chicago Clinics for an exploratory laminectomy. However, she shortly developed measles, thus delaying the operation. On August 10, 1933, examination revealed the dimple previously noted just above the spine of the 5th thoracic vertebra. This was not tender, seemed freely movable on the underlying tissues and no discharge could be expressed from it. Neurological examination revealed no abnormality of any of the cranial nerves. The tendon reflexes of the upper extremity were normal; those of the lower very brisk

the right somewhat more so than the left. A sustained ankle clonus was present on the right side and a transient clonus on the left. The right abdominal reflexes were sluggish, the left brisk. Both plantar reflexes were extensor in type. No definite sensory level could be found, although at times one gained the impression that sensory perception was not as acute over that portion of the body below the upper thoracic region. The finer modalities of sensation (two point discrimination, position-sense, stereognosis, &c.) could not be tested owing to the patient's failure to co-operate. There was no localized atrophy in any muscle group, but in general the strength was poor. No increased resistance was encountered on passive movement of the upper extremities, but in the lower extremities there was definite extensor rigidity, which was more marked on the right side. Co-ordination was fairly good throughout. A scoliosis with its convexity to the right in the upper thoracic region and a compensatory curve in the lower thoracic and upper lumbar region were present. The gait was spastic and the patient walked with small, uncertain steps. When standing with her feet together and the eyes open she did not sway, but on closing her eyes she tended to fall to one or other side.

A lumbar puncture at this time showed clear cerebrospinal fluid under a pressure of 110 mm. of fluid. Jugular compression (bilateral) caused a slow rise to 240 mm. and an equally slow return to 110 after release of the pressure, indicating a partial block of the subarachnoid space. Removal of 3 c.c. of fluid caused the pressure to fall to 80 mm. No cells were found in the fluid and the total protein content was 64 mgm. per 100 c.c. Roentgenograms revealed the defective closure of the laminæ of the 4th thoracic vertebra previously mentioned.

On August 16, 1933, under ether anæsthesia, a laminectomy of the 3rd, 4th and 5th thoracic vertebræ was performed. The spine of the 4th thoracic vertebra was absent and the imperfect closure of the posterior neural arch which had been observed in the roentgenograms was seen. The dimple was found to be continuous with a tough stalk of tissue about 3 mm. in diameter. This stalk contained a minute central canal from which thick caseous material could be expressed. The stalk extended from the dimple downward through the posterior spinal muscles, through the defect in the laminæ of the 4th thoracic vertebra to the dura mater, to which it was firmly attached. Beneath the dura mater at this point a firm mass was palpable. The dura mater above the tumour mass pulsated normally, but no pulsation was present below it. The dura mater was incised around the fibrous mass, exposing a thickened, white, and opaque arachnoid membrane. The arachnoid membrane about the margins of the tumour fused with the greatly thickened dura mater, and both were incorporated in the thick fibrous mass which was firmly adherent to the posterior surface of the spinal cord (fig. 1). This mass could be removed only by cutting it off the posterior surface of the spinal cord. This procedure revealed within the mass numerous cystic spaces containing a thick caseous material which on examination was found to contain many pus cells, but to be sterile on culture. After

removal of the fibrous mass palpation disclosed that the entire spinal cord was exceedingly firm at this level for a distance of 1·5 to 2 cm. Laterally, the spinal cord was firmly adherent to the much thickened arachnoid and dura mater, but no additional masses or abscesses were found. The dura mater was closed above the site from which the tumour mass had been removed, but closure at that point was not possible because of the large dural defect. The closure of the wound was made in the usual manner except that a small soft rubber drain was left in place.

The post-operative course was uneventful. Except for a moderate amount of pain at the operative site, the patient made no complaint. Both sensation and movement in the lower extremities remained good. Since the cultures of

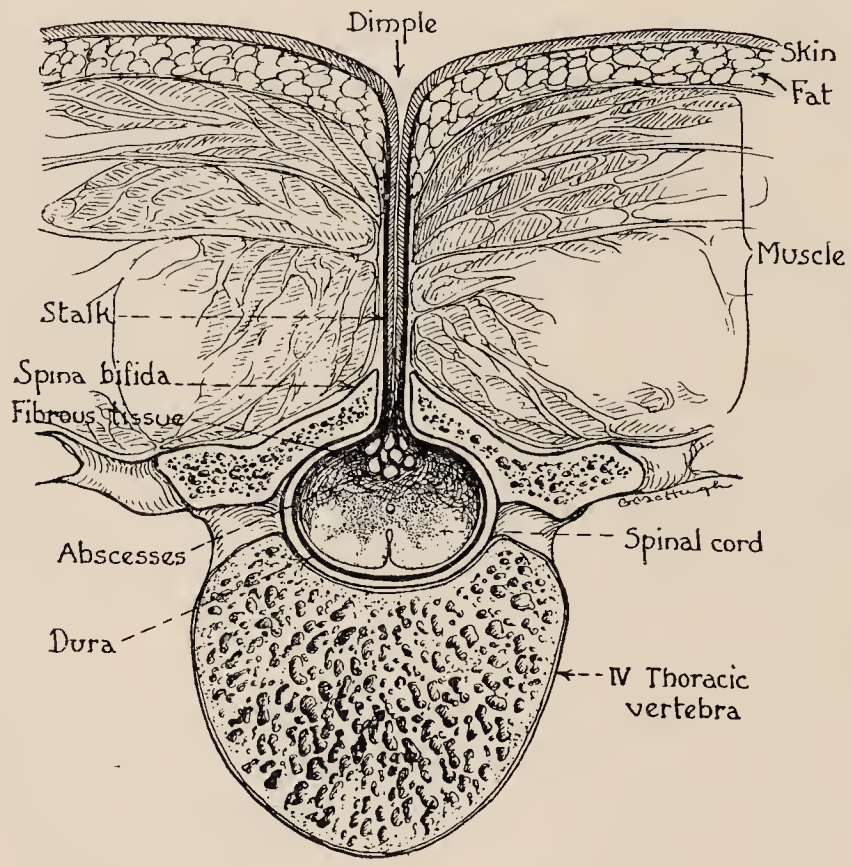

FIG. 1 (*Case* 1).—Diagrammatic representation of the congenital dermal sinus and the spina bifida of the fourth thoracic vertebra.

the purulent material were negative, the drain was removed on the second day and the wound healed per primam. For the first week she had urinary and fæcal incontinence, but then recovered complete control of her bowel, and partial control of micturition.

Neurological examination two weeks after operation showed little change from the findings on admission. An indefinite level of impaired perception of pin-prick was present at the 3rd thoracic dermatome. The reflexes of the lower extremities were still very brisk, with ankle and patellar clonus and bilateral extensor plantar responses. The strength in the lower extremities was fairly good, although at first the patient walked on a wide base with a spastic gait. In Romberg's position she was fairly steady even with the eyes closed.

She was discharged to a convalescent home and when seen six weeks after operation had no subjective complaints. She walked much better, although the

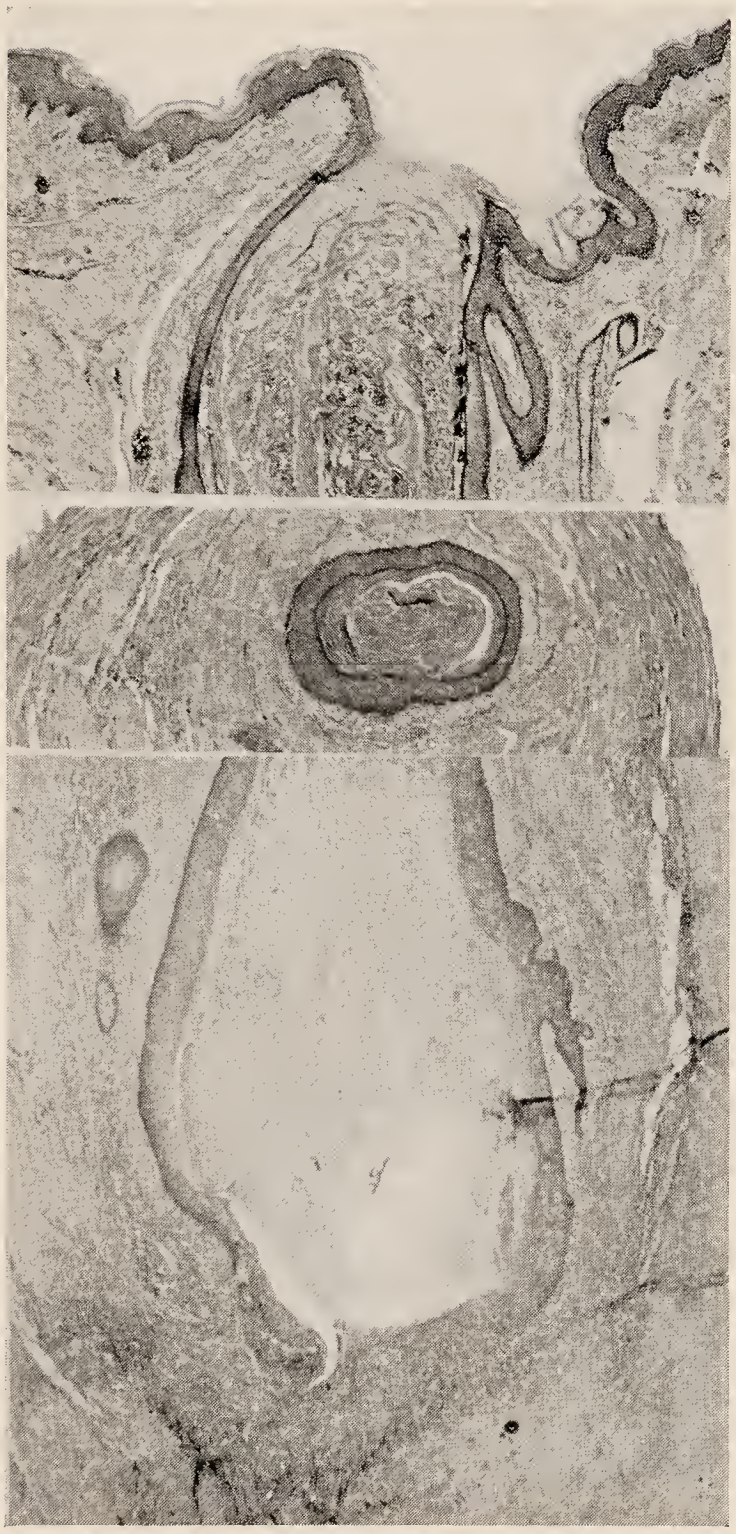

FIG. 2.

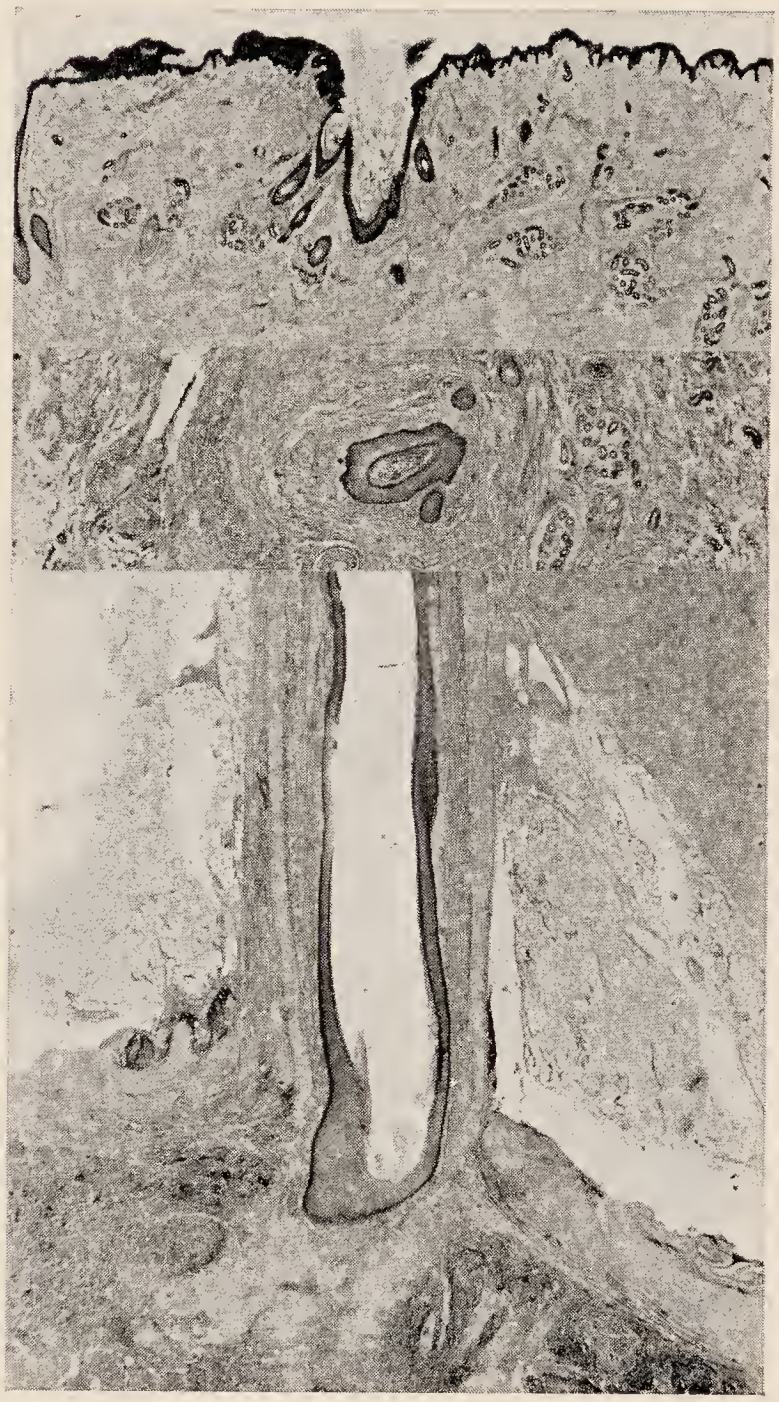

FIG. 4.

FIG. 2 (*Case* 1).—Composite plate of the congenital dermal sinus showing the stoma, the duct and a part of the abscess at the base of the sinus. Hæmatoxylin and eosin.

FIG. 4. (*Case* 2).—Composite plate of the dermal sinus showing the stoma, the epithelial tube, and its ending in an inflammatory mass beneath the dura. Hæmatoxylin and eosin.

To illustrate paper by A. Earl Walker and Paul C. Bucy.

gait was still somewhat spastic. She had perfect control over her bladder and bowel. The reflexes of the lower extremities were still quite brisk with ankle clonus and Babinski's sign on both sides.

On December 16, 1933, she was able to walk and run about without any sign of spasticity or limp. The tendon reflexes of the lower extremities were brisk and both plantars were extensor. A sustained ankle clonus was present on the right side. No sensory change could be demonstrated in any part of the body.

The family history is irrelevant except that a maternal cousin of the patient has a tuft of hair and an area of thin skin about 2 cm. in diameter in the upper lumbar region. The patient's father is suffering from schizophrenia of the paranoid type.

Pathological examination (Plate XIII, fig. 2).—The specimen removed at operation consisted of three pieces of tissue. The first was a bit of skin about 3 cm. in diameter, in the middle of the undersurface of which could be seen a round white tube of fibrous tissue with a small central orifice. The second was the stalk connecting the skin with the contents of the spinal canal. The third was a tough fibrous mass of tissue, cylindrical in form, 1·5 to 2 cm. in diameter, one surface of which was covered with dura mater while from the other thick caseous material could be expressed. The tissue was fixed in formalin, embedded in paraffin, and cut in serial sections, at right angles to the skin surface in the case of the first specimen and parallel to the long axis of the subdural mass. Cross sections were made of the stalk.

The sections were stained with hematoxylin and eosin for cellular detail; with van Gieson and Perdrau's method to show connective tissue. Sections from the subdural mass were stained by Loyez's method for myelin.

Study of these sections shows that the malformation consists of a tube of epithelial tissue opening out on to the surface of the skin at the dimple and extending downward to the mass lying beneath the dura mater, which was in turn closely attached to the posterior surface of the spinal cord. The skin about the mouth of the tube shows little change except some subdermal fibrosis. Near the stoma an occasional hair follicle is seen arising from the epithelium of the tube. Deeper, however, no hair follicles or other dermal appendages are present. The stoma itself and the canal of the stalk are lined with stratified squamous epithelium and plugged with desquamated epithelium arranged in concentric layers, in the centre of which is more debris irregularly arranged.

The epithelial lining of the tube becomes thinner as one approaches the inner end. The internal surface is devoid of papillæ and hence smooth. Although the cells of the stratum granolosum are less numerous, granules of keratohyalin are prominent. At the very bottom of the tube the epithelium is defective. Surrounding the tube is a thick layer of collagenous fibres which may be followed to the base of the tube where it is continuous with the thickened dura. Along the epithelial wall of the duct are small collections of lymphocytes and occasionally a calcospherite, both becoming more numerous as the stalk approaches the dura. The epithelial tube, however, terminates above the

dura, about 1½ cm. above the spinal cord, giving way to a mass of fibrous tissue. Beginning immediately below the lower and defective end of the epithelial tube, this fibrous mass becomes densely infiltrated with inflammatory cells, small round cells, plasma cells, macrophages and an occasional polymorphonuclear leucocyte. The dense connective tissue mass is composed largely of thick fibres of collagen, a few fine fibres of reticulin and a few fibroblasts. In many places within this thick felt-work of connective tissue small abscesses are seen containing cells similar to those noted above, and many multinucleated giant cells of the "foreign-body" type. No bacteria were seen in these abscesses and it will be recalled that no organisms could be cultivated from this purulent material. Nowhere is any nervous tissue seen.

Dr. George W. Barthelmez, of our Department of Anatomy, very kindly examined sections of this malformation. He was of the opinion that the histological appearance of the skin in the sinus with a narrow malpighian layer and no papillæ was embryonic in character and resembled that on the dorsal side of the finger tip in a four months' fœtus. He considered that the small epithelial proliferations from the skin of the sinus near its base were primordia of sebaceous glands, and comparable to findings in embryonic skin.

In brief, the pathological picture is that of an epithelial tube extending from the skin through unfused spinal laminæ to an inflammatory mass composed of numerous small sterile abscesses lying beneath a thickened dura, involving the arachnoid and attached to the posterior surface of the spinal cord. In this case of spina bifida, instead of the usual type of developmental abnormality in which a "firm fibrous band of connective tissue, fat, elastic tissue and striated muscular fibres passes through the vertebral cleft and connects the cord with the overlying skin" (Frazier [2]), the cord was connected by an epithelial tube passing through the defect in the lamina to the skin on the surface of the body.

This congenital malformation is identical with the pilonidal sinuses which occur in the sacral and coccygeal regions. The name pilonidal is a misnomer since only 40 per cent. of those sinuses contain hair (Owen [6]). For that reason we have called these malformations congenital dermal sinuses as we believe this more accurately describes the anatomico-pathological condition. Histologically, however, there is no distinction between the two anomalies.

With an understanding of the pathology of the case it is easy to explain the pathogenesis of the patient's earlier illness, diagnosed as tuberculous meningitis, from which she recovered. In the course of time infectious material undoubtedly found its way into the epithelial tube through the open stoma which appeared as an inoffensive dimple on the surface. This set up an inflammatory reaction within the tube

as is attested by the infection about the dimple and the sanguinous purulent discharge observed by the parents. This infection spread through the end of the tube to the dura, which was one with the connective tissue wall of the tube. The intradural abscess progressively grew, irritating the underlying leptomeninx and thus setting up a sterile meningeal reaction which was diagnosed as a "tuberculous meningitis." This not surprising error was reinforced by the unexplained death of the guinea-pigs.

If now in retrospect we continue the interpretation of this patient's story we must conclude that in the course of several months she overcame the localized infection with the resultant subsidence of the generalized meningeal reaction. She then enjoyed three years of apparent good health, at the end of which time sufficient infectious material again entered the epithelial tube to initiate a second illness. Again, we see evidence of extension of the inflammatory reaction, probably sterile, far beyond the seat of the infection as indicated by the paræsthesiæ of the ulnar surfaces of the fore arms (eighth cervical and first thoracic dermatomes). The proliferative process which succeeded the initial infection during the first years of life had undoubtedly left a thickening of the meninges, to which this second infection now added abscesses and inflammation sufficiently increasing the size of the connective tissue mass to compress the spinal cord. In addition, adherence of the inflammatory mass to the spinal cord produced a reaction within that structure. From both these factors a paraplegia resulted, but the sensory pathways were not affected. This resistance of the sensory fibres to various pathological processes which damage the motor tracts is, of course, a commonly observed phenomenon in many diseases of the spinal cord.

When our findings in this first case were reported to Dr. Joseph Brennemann of the Children's Memorial Hospital, Chicago, he immediately informed us that he had another young patient which he felt sure must be suffering from the same condition. Accordingly the following case was referred to us.

Case 2.—J. S., a 3-year-old white girl, was admitted to the University of Chicago Clinics from the Chicago Children's Memorial Hospital on March 9, 1934, with the complaint of stiffness and pain in the back and progressive weakness in both legs since June of 1933. She was the younger child in a family of two. Her mother had had two miscarriages. Her birth and developmental history were quite normal. A small hairy mole over the fourth lumbar spine had been noted since birth. She had had no illness or operations, but

in February of 1933 she fell from a chair and fractured her left femur, for which she was put in traction for five and a half weeks. During that time she complained occasionally of pain in the back, but it was not noticed until three months later that she had difficulty in walking upstairs and was unable to bend forward. Later, in August, she complained of itching and painful feet, and in September she was admitted to the Children's Memorial Hospital. Blood and urine examinations at that time were normal, the Wassermann and Kahn reactions on the blood were negative, and both Mantoux and Von Pirquet skin tests were negative. Spinal puncture on two occasions showed 700 and 680 cells, 83 and 32 per cent. respectively of which were lymphocytes. Culture of the first grew *B. proteus*, but no growth could be obtained from the second specimen. Tuberculous meningitis was suggested as the most probable diagnosis. Roentgenograms of the heart and chest were normal.

The child was removed from the hospital against the advice of the physicians, but returned because of weakness of the left leg. On January 3, 1934, the patient complained bitterly when the legs or feet were moved, and seemed to have paræsthesiæ in the left foot. Neurological examination showed that all the cranial nerves were normal; knee and ankle jerks on both sides were absent; both plantar reflexes were flexor. There was a marked weakness of the peroneal groups of muscles of the left leg, but the remainder of the muscles of the lower extremity seemed normal. No gross sensory change could be detected. At that time attention had been called to our first case, and the reddened lumbar nævus was then examined with great interest and the correct diagnosis was readily agreed upon.

On account of an outbreak of exanthemata, operation had to be postponed and the child was sent home until the period of quarantine was completed. During that time she seemed quite active and had no complaints until one week before admission to the University of Chicago Clinics, when the left leg became paralysed and the right very weak. Two days later she had incontinence of fæces and retention of urine with overflow incontinence. She had no appetite and was restless and irritable. On several occasions the hairy mole in the lower lumbar region became red and swollen.

Physical examination revealed a pale, poorly-nourished, but fairly well-developed girl of about $3\frac{1}{2}$ years of age lying quietly in bed, her left leg in the posterior half of a cast. She co-operated readily, but was apprehensive. Her general physical examination was negative except for the neurological findings.

The cranial nerves were unaffected and the upper extremities were normal in motor, sensory and reflex muscular status. The abdominal reflexes were absent. There was no movement of the umbilicus when the head was raised. At the first examination, March 12, 1934, both ankle-jerks could be obtained, but four days later both were absent. The knee-jerks were constantly absent. The plantar reflexes were bilaterally flexor. No clonus could be elicited at either knee or ankle. No sensory change over any part of the lower extremities could be elicited. The left leg was completely paralysed except for slight movement at the ankle and toes. The right leg could be flexed at the hip and

knee, but not extended. The ankle could be both flexed and extended, but plantar flexion was much stronger than dorsi-flexion. The toes could be readily flexed, but extension was weak. There was no localized atrophy of the leg muscles, although all muscle groups were small. The right leg was flaccid and the left moderately spastic.

No change in any modality of sensation could be detected. There was a moderate rigidity of the neck and marked stiffness of the spinal column so that it could not be bent forward. Over the fourth lumbar spine a red hairy nævus about 1·5 cm. in diameter was present, in the centre of which was a minute crust. This entire area was rather firmly attached to the underlying tissues, and was tender to palpation. There was no tumefaction and no palpable spina bifida.

The child was able to sit up alone but unable to maintain that posture unless her hands were kept on the bed behind her to prevent falling backward. She was unable to stand or walk. She had urinary retention necessitating catheterization three or four times daily and enemas were necessary for the evacuation of her bowels.

Roentgenograms taken of the pelvis and lower part of the vertebral column showed incomplete fusion of the laminæ of the sacral segments. Blood examinations were negative except for a leucocytosis of 13,600 and 24,200. Urine analysis showed many leucocytes in the sediment but was otherwise normal. During her pre-operative period she had no fever. A lumbar puncture was made between the 5th lumbar and the first sacral spine and thick caseous pus encountered. A syringe attached to the needle aspirated 2 or 3 c.c. of a dirty fluid containing many particles of debris. A culture of this fluid grew *B. coli*.

On March 17, 1934, under general ether anæsthesia, a midline incision was made from the 11th thoracic to the 1st sacral spine, the reddened nævus being excised. Extending from the centre of this area a small white fibrous tube was seen passing between the spines of the 4th and 5th lumbar vertebræ down to the dura to which it was firmly attached. The lamina of the 2nd, 3rd, 4th and 5th lumbar vertebræ were removed. The bone appeared normal. The dura, however, was tense and bluish, and there was a mass at the point where the fibrous tube entered the meninges. The dura, firmly adherent to the arachnoid, was incised at this point and the incision carried through 1·5 mm. of reddish granulation tissue. A cystic cavity 4 to 5 mm. in diameter filled with thick creamy pus and some sebaceous material was encountered. The incision was then extended downward about the mass which lay between the 4th and 5th lumbar vertebræ. Below that, the dura was readily separable from the arachnoid, beneath which the roots of the cauda equina could be seen imbedded in a greyish gelatinous material. The abscess wall, however, did not extend below the level of the 5th lumbar vertebra and the arachnoid was not opened below that point (fig. 3). The skin and fibrous tube were cut off the posterior surface of the arachnoid and pieces were taken from the wall of the abscess for further microscropic study.

In order to reach the upper end of the abscess, the 1st lumbar and 12th thoracic vertebral laminæ were removed. After incising the dura beneath the 12th thoracic laminæ the posterior surface of the spinal cord was seen. The abscess was then opened up to a point where the cavity seemed to continue downward in a hole which appeared to sink into the dorsal surface of the spinal cord. It was not followed further for fear of injuring the substance of the spinal cord.

The dura was left open and two small soft rubber drains were placed in the abscess cavity and led out from each end of the wound. The muscles and fascia were closed with interrupted catgut and the skin with silk.

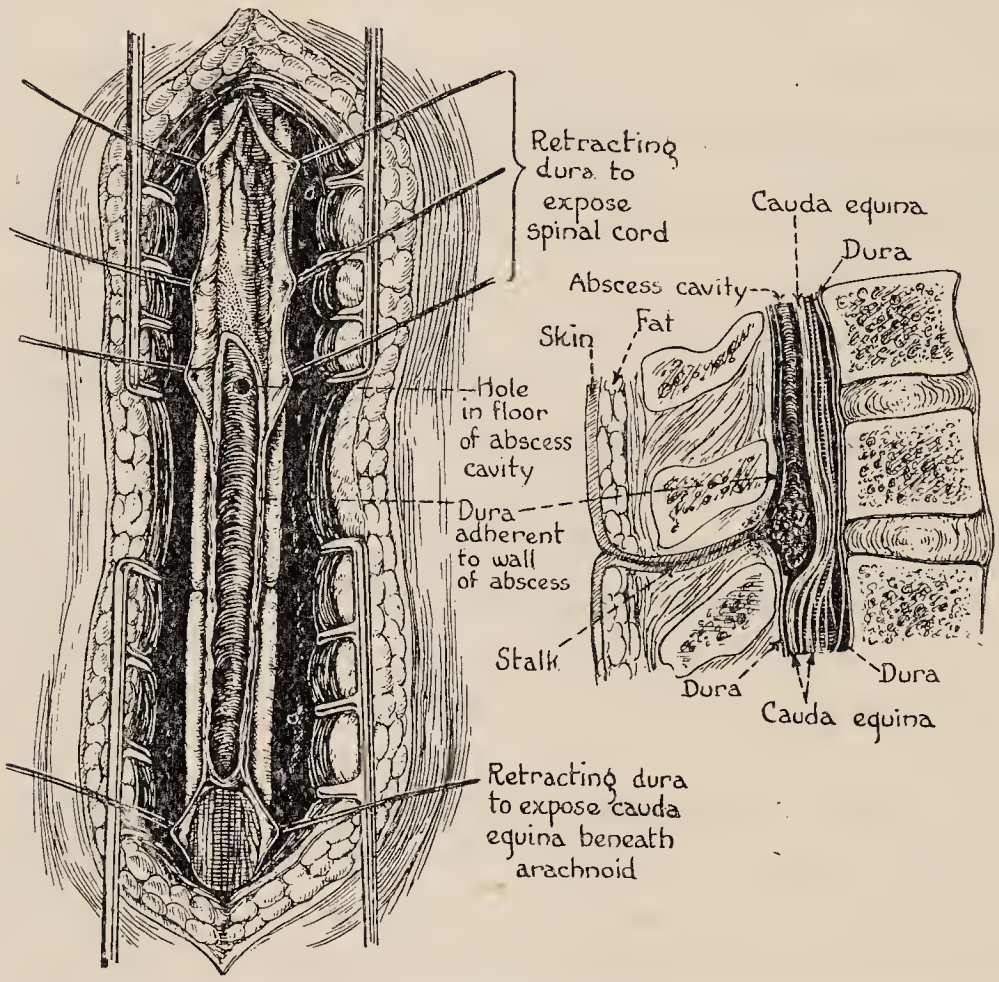

FIG. 3 (*Case* 2).—Semi-diagrammatic illustration of the operative findings in Case 2. The situation and relation of the meninges and cauda equina to the abscess are shown.

The patient had a slow but uneventful convalescence. Both drains were removed four days after operation, and very little drainage occurred following this. Two weeks after operation the wound was completely healed. Movement in both legs increased, although the right continued to be stronger. Four weeks after operation the patient was able to sit up unassisted, but could not stand even with help. No sensory disturbance could be detected for pain, touch, position or vibration sense. In the right leg the patient had fairly good movement of all muscle groups, but flexion of the left knee was weak, and flexion, inversion and eversion of the left ankle were extremely weak, there being just slight motion against gravity. The right leg was flaccid, the left spastic and painful on movement. Both knee-jerks and the right ankle-jerk were absent, but the left ankle-jerk was greatly exaggerated with a sustained clonus.

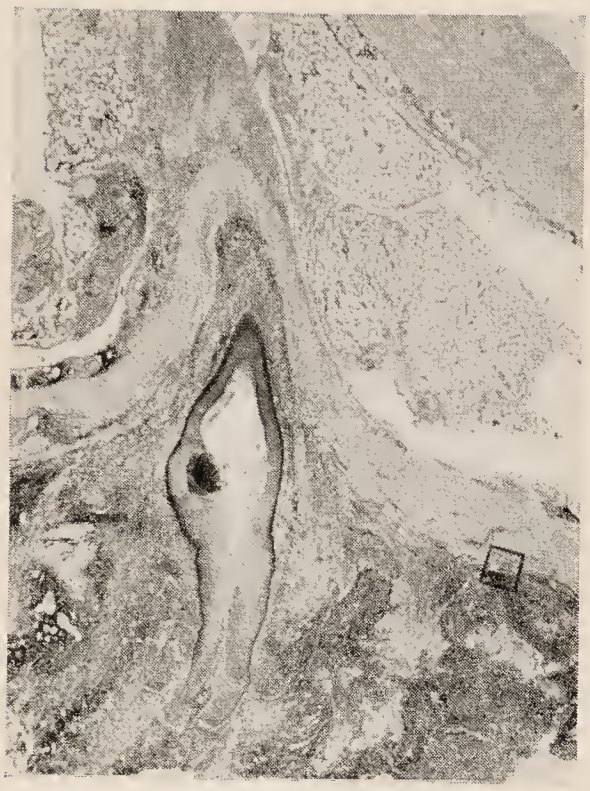

FIG. 5.

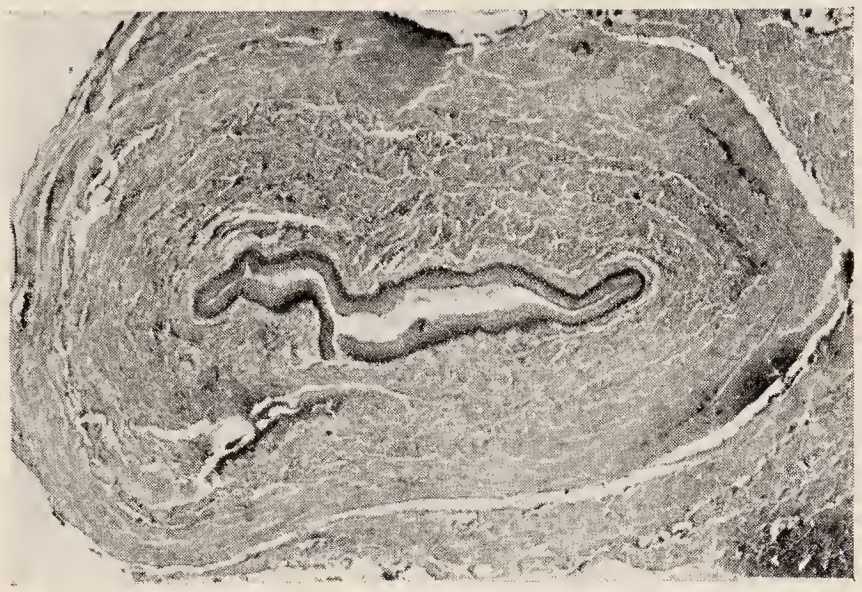

FIG. 7.

FIG. 6.

FIG. 5 (*Case* 2).—Photomicrograph showing the lower end of the dermal sinus penetrating the dura and opening into an inflammatory mass. Hæmatoxylin and eosin.

FIG. 6 (*Case* 2).—A higher power photomicrograph of the small square marked in fig. 5. The nature of the inflammatory reaction is well shown, and its extension into the dura mater is shown by the clumps of cells, mainly small round cells, in the dura mater in the upper half of the illustration. Hæmatoxylin and eosin.

FIG. 7 (*Case* 3).—Cross section of the dermal sinus which led from the skin to the meninges in Case 3. Hæmatoxylin and eosin.

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When last seen on May 29, 1934, she was able to walk without assistance, although there was some external rotation and eversion of the foot. All movements of the foot and leg are now possible. She felt quite well and was very lively.

Pathological examination (Plate XIII, fig. 4).—The skin containing the dimple visible on the surface and the subdural mass leading down from it were sectioned serially and stained with hæmatoxylin and eosin, van Gieson's acid picro-fuchsin and impregnated by Perdrau's method.

Sections of the skin revealed a normal surface epithelium, the basilar layer of which contains a brownish pigment. There were the usual papillæ and in the dermis are found many hair follicles, sebaceous and sudorific glands, all apparently perfectly normal. In about the centre of the section is seen an invagination of the epidermis forming a narrow epithelial tube which passes downward into the deeper layers of the skin. The epithelial lining of this tube is very similar to the surface epithelium except that the pigmentation of the basal layer and the normal papillæ are absent. There are a few fine hairs which project from the neighbouring dermal connective tissue through this epithelium into the tube. There are also a few sudorific and sebaceous glands connected with this tube. The lumen is filled with cornified, desquamated epithelial debris. In the connective tissue of the dermis immediately about the tube is a rather marked cellular infiltration. The cells are largely of the lymphocytic series.

Sections of the subdural mass show the downward extension of this epithelial tube. It is similar to that near the orifice on the skin surface except that there are no hair follicles and no glands. Here again there are no papillæ. The tube passes beneath the dura mater, which is continuous with the dense connective tissue sheath of the epithelial tube. The tube terminates a short distance beneath the dura mater, its lower end being open and its lumen communicating with an extensive subdural abscess (Plate XIV, fig. 5).

The subdural abscess is composed of a mass of inflammatory cells of which about half are polymorphonuclear leucocytes and half lymphocytes. There are in addition a few endothelioid cells and a few multinucleated giant cells. These cells are enmeshed in a loose reticulin network. The abscess is intimately connected with the dura mater, strands of which pass into the inflammatory mass (Plate XIV, fig. 6). The remainder of the dura mater is infiltrated by small round cells which lie about the vessels and between the collagenous fibres. There is no epithelium within the abscess other than the extreme lower end of the epithelial tube mentioned above.

This patient's history was not unlike that of the first case. Both were diagnosed as tuberculous meningitis, and both had slowly progressing signs and symptoms of spinal cord involvement with mild remissions. Neither had significant sensory changes either before or after operation. The dermal sinus in this case, as in the first, consisted of an epithelial tube surrounded by collagenous fibrous tissue. However, unlike the first case, it contained dermal appendages and the sinus could

be traced down through the dura, whereas in the former case the sinus could be followed only to the epidural mass of inflammatory tissue. The abscess cavity was much more extensive than in the first case, and it was impossible to extirpate it completely. It is noteworthy that the sinus passed between intact vertebral arches, instead of through a defect in the laminæ as in the first case.

The production of an abscess in the subarachnoid space is unquestionably due to the same factors as in the first case—the presence of a long epithelial tube communicating with the skin which readily allows infecting organisms to reach the meninges.

The patient has been followed but a few months, but the improvement in this short time is promising.

Dr. Dallas B. Phemister informed us that some twelve years previously he had operated upon a similar case. He kindly had this patient attend to the out-patient department and secured for us the microscopical preparations of the material removed at operation as well as the entire record of the case. It is through Dr. Phemister's kindness that we are at liberty to include his case with ours in this Report.

Case 3.—T. S. A coloured girl, aged 3 years, was admitted to the Presbyterian Hospital, Chicago, October 21, 1921, on the service of Dr. D. B. Phemister. The patient was the second child in a family of two, born at full term spontaneously. Her development was quite normal, and the parents had noticed no abnormalities except that in the lower lumbar region the patient had a dimple about which were a few hairs. This had been present since birth. At 14 months of age the child had pneumonia, and about the same time a family physician had pulled some hair from the skin surrounding the dimple. Following this it discharged pus for a short time and then healed. Since then it had swollen from time to time, broken down, discharging pus for a period, and then healed again. The area was constantly tender, and the patient complained of severe pain in the back and occasional occipital headaches.

Examination revealed a fairly well nourished young negro girl presenting typical racial characteristics. She was not acutely ill. Physical examination at that time was quite negative except for the local findings. At the level of the spine of the 5th lumbar vertebra in the midline was a hairy, pigmented area in the centre of which a minute hiatus could be detected. The skin was tender on palpation. No neurological examination was recorded. The child walked with the knees semiflexed.

Laboratory examination: The urine and blood were normal, except that there was a leucocytosis of 17,000. Roentgenograms of the spine showed a lack of fusion of the spinous process of the last lumbar and first sacral vertebræ.

On October 25, 1921, the patient was operated upon under ether anæsthesia by Dr. Phemister. An incision about 6 cm. long was made parallel with the vertebral spines over the last lumbar and sacral vertebræ. The hairy pigmented area surrounding the minute opening in the skin was excised. A probe put into the hiatus was seen to pass down a stout fibrous tube through a defect in the spinous processes of the 5th lumbar vertebra, and to rest upon the spinal meninges. In removing the fistulous tract the meninges were opened and about six drops of glairy, gelatinous, slightly purulent material escaped through the rent. In order completely to remove the sac a part of the dura had to be cut away. Within this, what appeared to be a friable sac wall was dissected free from the nerve roots. The dura was left open and cerebrospinal fluid escaped freely. The fascia was closed with catgut and the skin with silk. One plain gauze was left in the wound and removed the following day. On October 27, 1921, two stitches were removed and a few cubic centimetres of pus expressed. For a week dark-brown pus drained freely from the wound, and then the discharge gradually decreased and the wound granulated, so that on the patient's discharge on November 15, 1921, it had completely healed.

The patient was seen again on March 2, 1934. Her mother stated that following the operation in 1921 the patient's gait had rapidly improved, and that she had no bowel or bladder disturbance. At that time, March 2, 1934, she had no complaint, and her mother regarded her as normal. Examination showed that the operative scar was well healed with no swelling. There was no sensory loss in any part of the body, no motor weakness, and the gait was normal. The knee and ankle jerks on both sides were absent.

Pathological examination.—Through the kindness of the pathological laboratory of the Presbyterian Hospital, a slide containing three pieces of tissue was examined. The first specimen is a cross-section of heavily pigmented epidermis and subcutaneous connective tissue. Numerous dermal appendages, sweat and sebaceous glands and hair follicles are present, and present no abnormality. There is a small irregular depression in the centre of the epidermis containing cornified epithelial debris on one side and the opening of a hair follicle on the other, possibly the stoma of the dermal sinus. The second piece of tissue is a cross-section of a small tube of stratified squamous epithelium showing no pigment in the basal layers. It is surrounded by a thick layer of collagenous connective tissue and outside of this is considerable adipose tissue. The lumen of the tube contains a small amount of cornified epithelial debris. The inner surface of the epidermis is devoid of papillæ, and no dermal appendages are seen. Occasional small round cells are scattered throughout the connective tissue surrounding the epithelial tube (Plate XIV, fig. 7). The third section is a fibrous and collagenous connective tissue studded with small abscesses. These are composed of masses of small round cells and phagocytic cells. In other areas small collections of these cells are present without any evidence of central necrosis. A few polymorphonuclear cells are present. At one corner of this specimen a small piece of epidermis

identical with that seen in the first piece of tissue is present, and in the opposite corner an arc of epithelial tissue, probably a small segment of a hair follicle.

Even with these few sections it is evident that the condition present in this case is very similar to that in the first two patients. The extent of the dermal sinus is unknown, but as demonstrated grossly it probably extended at least to the subdural inflammatory mass.

As in the previous cases an epithelial tube leading from the skin to an inflammatory mass beneath the dura mater was found at operation. The histopathology of this was similar to that of the specimens from the first two cases. However, it is interesting to note that there was no pigment in the basal layers of the epithelium of the tube, such as was present in abundance in the normal epidermis. The same phenomenon was noted in Case 2 in which the surface epithelium, though from a white girl, was heavily pigmented, whereas that from the tube contained no pigment.

Dr. Samuel C. Harvey, of the Yale University School of Medicine, has also informed us that several years ago a similar case was operated upon in his clinic. This case was recorded by Moise [5] and is related here in detail.

Case 4.—A white male, aged 18, was admitted to the New Haven Hospital on September 10, 1924. He had always had in the lower lumbar region a sinus discharging a watery fluid at short intervals. A week before admission the skin about the sinus became tender, and two or three days later his back began to ache. The day before admission he had headache, general malaise and anorexia. There was no nausea, vomiting or convulsions. When admitted his temperature was 101·8° F., pulse 86, and respirations 20 per minute. He appeared ill, his face was flushed, and his expression anxious. The heart, lungs and abdomen were normal. Neurological examination showed a very stiff neck, normal tendon reflexes in the upper extremity, but absent knee and ankle jerks. Kernig's sign was present. In the midline over the upper sacral region was an area of red and tender skin surrounding a small sinus from which some thin pus could be expressed.

A roentgenogram of the sacrum showed a sacralization of the 5th lumbar vertebra, an irregularity in the fusion of the spines of the 5th lumbar and the 1st sacral vertebræ, and a flattening of the spine of the 1st sacral segment with a defect below this level.

Lumbar puncture provided a cloudy cerebrospinal fluid under increased pressure containing 1,450 cells per cubic millimetre, predominately polymorphonuclear leucocytes. A few Gram-negative cocci were seen in smears, and culture showed hæmolytic *Staphylococcus albus*. Daily lumbar puncture failed to improve his condition, so operation to remove the sinus was advised.

Operative note: September 23, 1924. The sinus was injected with methylene blue and excised with the surrounding tissue. The sinus extended through a small bony defect, measuring 1 cm. in diameter, just to the right of the midline at the junction of the 1st and 2nd sacral vertebræ. The incision was then extended and a laminectomy performed. The spine of the 1st sacral vertebra was flat and was removed. The underlying dura was stained deeply with methylene blue. There was a tuft of granulation tissue just beneath the defect in the spinal column. This was excised after the dura had been opened. A small rubber tissue drain was inserted through the upper part of the incision down to the dura. The wound was closed in layers.

Histological examination of the excised sinus showed it to be a tube lined with several layers of stratified squamous epithelium surrounded by a dense fibrous wall.

The patient rapidly improved following operation, although for several days he suffered severe pains in his back and legs. Spinal fluid drained from the wound for nine days. At the time of discharge the patient felt quite well, and there were no neurological abnormalities.

The author considered this case an example of pilonidal sinus which, instead of ending blindly, communicated with the subarachnoid space through a defect in the spines of the sacral vertebræ. It is evident that this is similar to the previously mentioned cases, and the only unusual finding is the direct opening of the tube into the subarachnoid space rather than into an inflammatory mass about the meninges. It is likely, however, that this communication was secondary to the infection and was not present in the original congenital defect.

A careful review of the literature has revealed three additional cases. A case almost identical with that described by Moise was reported by Ripley and Thompson [1] in 1928.

Case 5.—D. L., male, aged $3\frac{1}{2}$ months. The patient had a normal birth. For two weeks prior to admission he had been fretful, screaming when lifted. On admission the child had a temperature of 101·4° F., pulse 148, respirations 48. Physical examination revealed consolidation of the right upper lobe of the lung. Four days later, although the child's condition was slightly improved, pus was noted at the bottom of a sacro-coccygeal dimple, and a probe was slipped in 4 cm. without encountering resistance. On lumbar puncture 2 cm. of thick yellowish pus was removed by aspiration. Examination of a smear of the material showed *Staphylococcus albus*. Operation was then performed, excising the sinus as high as the lumbo-sacral junction. One part of the sinus lay within the sacral canal. The second sacral arch was absent. A drain was inserted and the wound closed.

When opened the sac removed at operation was found to contain sebaceous material and a hair. Microscopical study of the sinus showed it to be of the same structure as normal skin with an inflammatory reaction about it.

The wound drained poorly and the patient's general condition became grave. Lumbar puncture was performed once or twice daily to drain the spinal fluid and relieve the intracranial tension. On the eighth day after operation the spinal fluid appeared clear but was under considerable pressure and contained 430 cells mainly polymorphonuclear leucocytes. The patient died on the ninth post-operative day. At autopsy methylene blue injected into the lumbar subarachnoid space was found in the operative wound. There was a very marked purulent exudate over the surface of the cerebral hemispheres.

This case is identical with that reported by Moise. Whether death was due to the defective drainage of the sinus, or to the failure of spinal fluid to escape from the wound, cannot be said.

The second case found in the literature is apparently similar to the first three of the above five cases, although there is no note as to the depth of the dermal sinus, and hair was found at its base. It was reported by W. and N. Sharpe [8] (pp. 269-371) as a case of intradural dermoid.

Case 6.—F. S., male, aged 3 years, was apparently a normal child except for a small pinhead-size depression over the spine in the lumbar region present since birth. His back was rigid, and from time to time a thick yellow discharge appeared at the site of the depression. Several operations superficially excising the sinus only left the patient with a marked weakness of both legs. The wound broke down and continued to drain purulent material.

At 3 years of age there was a spastic paralysis of both lower extremities but no sensory changes in any part of the body. Roentgenograms showed a defect in the left lamina of the 11th thoracic vertebra. A laminectomy was performed with excision of the fibrous sinus which was attached to an inflammatory mass about the posterior aspect of the dura. When this was opened around the nodule a mass of caseous material containing hair was found imbedded between the dura and the left side of the spinal cord. This was removed, a drain inserted and the wound closed.

The wound healed slowly, but otherwise the convalescence was uneventful. Motion returned in the legs in three weeks, and within a year he was able to walk well, although the left leg was dragged slightly.

No pathological description of the tissue removed is given, but it seems probable that this is another example of a dermal sinus extending downward through unfused lamina to end around the spinal cord. The striking point was the presence of hair at the base of the sinus, a phenomenon which is not unexpected since in the second case there was hair in the upper portion of the sinus.

All of the above six cases of congenital dermal sinuses were asso-

ciated with evidence of meningeal irritation and infection due to infected material having passed down the sinuses to these tissues. Clark [1] has reported a case which seems similar to these, although there was at no time any evidence of meningeal infection, nor was the nature of the lesion confirmed at either operation or necropsy.

Case 7.—L. N., female, aged 13 years. Her development was normal. It was noted that from birth the patient had a small depression in the median line between the 4th and 5th cervical spines out of which grew a small tuft of hair about half an inch in length. She lacked dexterity and speed in executing delicate movements with her fingers and hands, but there was no atrophy or motor weakness except a slight deficiency in adduction, abduction, flexion and extensisn of the little and ring fingers of both hands. The remainder of the neurological examination was quite negative.

Roentgenograms showed no defect in the 4th or 5th cervical vertebra but a cleft in the arch of the 3rd.

From the clinical description it is, of course, impossible to be certain of the exact nature of the lesion, but it is not unlikely that this is an example of the same malformation which has not yet become infected.

A careful search of the literature has failed to reveal further examples of this congenital malformation. The similarity of all seven cases, and the evident mode of infection through the dermal sinus, is quite striking and merits recognition as a clinico-pathological entity. The fact that two of these cases presented a clinical resemblance to tuberculous meningitis is still greater reason for the consideration of this congenital anomaly in any chronic meningitis, particularly as operation offers such an excellent result. The presence of the minute opening of the sinus in the skin, commonly referred to as a dimple, from which a discharge occurred at intervals, had been noted by the parents in all cases associated with meningeal irritation, before the patients were referred to a physician. This anomaly when accompanied by the typical history of fever, stiffness and tenderness of the back, neck rigidity, some degree of motor weakness in the lower extremities and the finding of a pleocytosis in the spinal fluid, should suffice to make possible the diagnosis of infected congenital dermal sinus extending to the spinal meninges.

Association with spina bifida.—It will be noted that every case was associated with a defect in the posterior neural arch which could be demonstrated radiographically. However, the defect demonstrable in the second case is the normal state for the sacral vertebræ at this age and cannot be considered as having any association with the congenital

dermal sinus which, unlike the condition found in the other cases, passed through the opening normally present between the laminæ of the 4th and 5th lumbar vertebræ. Thus, although it can be stated that this peculiar malformation is usually associated with a spina; bifida it is not necessarily so.

Although spina bifida occulta is generally considered as of not infrequent occurrence, there are very few statistics on which to base this conclusion. Mayer [4] states that a complete splitting of the sacrum occurred in 3 per cent. of cases, and that in children over 10 years of age the first sacral lamina was not fused in 24 per cent. of cases in the general population. These figures indicate a much higher incidence of spina bifida than that reported by Sutherland [9] at the Mayo Clinic. In approximately 12,000 roentgenograms of the spine he found 621 cases of spina bifida, an incidence of slightly more than 5 per cent. The condition was twice as common in males as in females. It involved the 1st and 2nd sacral vertebræ in 70 per cent. of cases, the 5th lumbar in 24·5 per cent. No mention is made of the incidence of spina bifida occulta in the thoracic region, but four cases are presented with the defect in that region.

In 1920 Theodora Wheeler reviewed the literature to that time, and in a group of 1,000 consecutive X-ray plates of the sacrum found the incidence of spina bifida occulta of the first sacral vertebra to be 13·1 per cent., and the laminæ of the entire sacrum were unfused in 2·89 per cent. of cases. The 5th lumbar was found unfused in 23 cases of the 1,000, and the atlas, the only other vertebra in that series with spina bifida, in 1·47 per cent.

Table I shows the incidence of spina bifida occulta in a series of roentgenograms of the spine taken at the University of Chicago Clinics in the past six years. These include films taken to show trachea, cervical spine, colon, chest, thoracic spine, lumbar spine, urinary tract, pelvis, sacro-iliac joints, and lumbo-sacral spine; some of which only incidentally showed the vertebræ. The total number of patients of whom roentgenograms of these parts were taken was noted, and then twenty representative roentgenograms of each studied to determine the mean number of vertebræ included in any roentgenogram of any of the above structures. From these data it was a simple mathematical problem to calculate the number of times each vertebra had been shown in the total number of roentgenograms. All cases of spina bifida occulta were then collected and reviewed to determine the location of the anomaly, its association with enuresis, motor or sensory defects, local

manifestations such as hirsuties or lipoma, and other congenital abnormalities. These data are given in tabulated form in Table II. In all 123 cases of spina bifida occulta, of which seventy-five were in

TABLE I.

Vertebra	No.	Total number Roentgenograms	No. of spina bifida	Percentage
Cervical ..	1	593	1	0·17
	2	735	0	0·00
	3	912	0	0·00
	4	1,149	0	0·00
	5	2,815	1	0·03
	6	4,088	1	0·02
	7	5,381	2	0·04
Thoracic ..	1	5,406	4	0·09
	2	4,989	5	0·10
	3	4,234	2	0·05
	4	2,182	4	0·18
	5	1,456	1	0·07
	6	1,500	1	0·07
	7	1,427	1	0·07
	8	1,770	1	0·06
	9	2,635	1	0·04
	10	3,584	1	0·03
	11	4,421	0	0·00
	12	5,967	1	0·016
Lumbar ..	1	6,290	1	0·015
	2	6,712	2	0·03
	3	6,594	2	0·03
	4	7,680	4	0·05
	5	7,516	28	0·37
Sacral ..	1	6,945	113	1·62
	2	6,301	41	0·65
	3	6,301	32	0·51

TABLE II.

	No. of cases	Enuresis	Sensory disturbance	Motor disturbance	Local manifestations (hair, dimple, &c.)	Other congenital abnormalities	No signs nor symptoms directly referable to lesion	Low back pain
Cervical	2	0	0	0	0	1	1	0
Thoracic	9	1	0	1	1	4	2	0
Lumbar	1	0	1	1	1	1	0	0
Lumbo-sacral (L_5-S_3)	111	16	3	5	11	16	78	42

males and forty-eight in females, were found. The most frequent site of the defect was in the lumbo-sacral region, 111 occurring there. Nine were present in the thoracic region, but in none except the first case

reported here was there any external manifestation of the defect, but four had scoliosis and other skeletal abnormalities. Three had several hemivertebræ and two of these had associated anomalies of the ribs.

Genesis of congenital dermal sinuses.—The consideration of the development of these congenital dermal sinuses leads to a discussion of the theories advanced for the ætiology of pilonidal sinus. In 1892 Mallory [3] came to the conclusion that these malformations were due to a failure of obliteration of the medullary canal in the coccygeal region. This view has found much support, the most recent coming from Weeder [11] in an article in 1933. It is obvious, however, that this theory will not explain the presence of these dermal sinuses in any

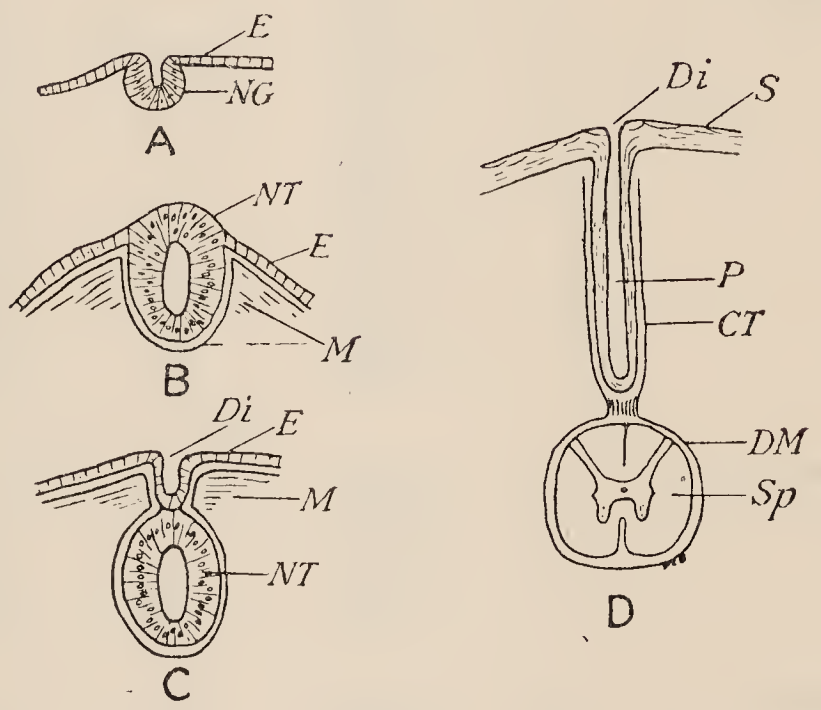

Fig. 8.—Sketch illustrating the possible development of the malformation : E., ectoderm ; N.G., neural groove ; N.T., neural tube ; M., mesoderm ; Di., dimple ; S., skin ; P., congenital dermal sinus ; C.T., connective tissue sheath of sinus ; D.M., dura mater ; Sp., spinal cord.

region of the body but the sacro-coccygeal, and that some other explanation must be given for their presence in the lumbar, dorsal and cervical regions.

The conclusion would seem justifiable that at the time in embryological development (the first month of intra-uterine life) when the cutaneous epithelial ectoderm and the neuro-epithelial ectoderm should have separated, the cleavage between them was incomplete at the particular point where the sinus occurred. Thus, the neural tube carried down with it a narrow invagination of the skin, clothing it with a mesodermal covering continuous with its own connective tissue covering, the meninges. The cause of this abnormal development remains one of the unexplained perversities of intra-uterine development. We have attempted to illustrate this interpretation of the development of this

condition in the accompanying sketch (fig. 8). Whether this same explanation of the ætiology is true for pilonidal sinuses cannot be stated, but it is at least a possibility (Stone [10]).

Summary.

1) Seven cases presenting a clinico-pathological entity are described. A congenital dermal sinus manifested externally by a small dimple in the midline of the back becoming infected, discharges periodically serous or purulent material. This sinus extends to the meninges and its lumen acts as a pathway for infection to reach these structures. When this occurs, producing abscess formation, fever develops, the back becomes painful and rigid, and the neck stiff. Weakness of the lower extremities ensues, but sensory disturbances are slight or absent. The spinal fluid, usually sterile, shows a pleocytosis.

(2) Surgical removal of the sinus and subdural abscess with drainage produces a complete cure in almost every case.

(3) The relation of this congenital dermal sinus to spine bifida occulta is discussed and the incidence of the latter in a series of 7,500 roentgenograms taken of the spine in a general medical and surgical clinic is given.

(4) The ætiology of these congenital dermal sinuses and pilonidal cysts is discussed.

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